COARCTATION OF THE AORTA, ANOMALOUS CORONARY ARTERY AND PATENT DUCTUS ARTERIOSUS*

REPORT OF CASE WITH SUBACUTE BACTERIAL ENDOCARDITIS, SHOWING MYCOTIC ANEURYSMS OF THE AORTA AND SUPERIOR MESENTERIC ARTERY

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THE diagnosis of coarctation of the aorta has been made more frequently since the appearance of Abbott's[1] and Blackford's[2] excellent articles. In 1928 Abbott was able to collect two hundred cases of coarctation of the aorta which were found at autopsy. Twenty-one of these were recognized clinically.

The signs of coarctation of the aorta are mainly vascular. The cardiac findings, which are often present, are an enlargement of the left ventricle (secondary to the hypertension in the upper body), and a systolic murmur along the left sternal border. This murmur is usually maximal in the back between the scapulae. The chief vascular finding is a change in tension and form of the pulse in the upper body as compared with the lower. In fact, Abbott states, "A diminished blood pressure in the lower extremities, with retardation and diminution or complete absence of the femoral pulse in the presence of a contrasting hypertension in the upper extremities, and a hard, full radial pulse is in itself ground for a provisional diagnosis." The second characteristic vascular sign is the evidence of collateral arterial circulation over the chest. Roentgenologic examination may show scalloped rib margins (a sequel of the hypertension and collateral circulation in the intercostal arteries), or abnormal pulsation of the ascending aorta, as compared with an undemonstrable or only slightly pulsating descending aorta.

Three times as many cases of coarctation are found in men as in women. Death is most apt to occur in adolescence or in early middle life. In their order of importance, death most commonly results from myocardial failure, rupture of the heart or aorta, cerebral hemorrhage, or mycotic endarteritis.

In the following case, coarctation was not recognized until a week before death.

REPORT OF CASE

W. S., a sixteen-year-old white, single, schoolboy entered the University of California Hospital on the Surgical Service on June 29, 1932, complaining of abdominal pain. When the patient was four years old he had an appendectomy with drainage because of a ruptured appendix. Since that time he had had occasional abdominal distress, accompanied by eructations of gas. When he was six years old he had measles complicated by otitis media, followed by intermittent attacks of otorrhea. When he was twelve years old, reddish urine was noted for a short time.

A year before his entry he was ill for two weeks with "pneumonia." Following this his abdominal symptoms increased in severity and he began to notice dyspnea and easy fatiguability on exertion. Two weeks before entry he was in bed several days with fever and generalized aching. He then began to have constant, aching, nonradiating epigastric pain, lasting five days. The pain was accompanied by nausea, but not by vomiting, until two days before entry, when he vomited, several hours afterward, the food eaten at breakfast. This was followed by severe, generalized abdominal colic which persisted to the time of entry. There was no constipation.

Physical Examination.—Physical examination at entry showed a pale, slender boy, who thrashed about in bed complaining of severe abdominal pain. The temperature, pulse rate, and respiratory rate were normal. The tongue was heavily coated. The tonsils were moderately enlarged and cryptic. The heart showed no signs of enlargement; the aortic second sound was accentuated; and a loud systolic murmur was heard over the precordium. The brachial blood pressure was 140 systolic, 80 diastolic. The abdomen was tense, but there was no distention, spasm or localized tenderness. Peristalsis was audible throughout.

An examination of his blood revealed 68 per cent hemoglobin (Sahli), 3,900,000 erythrocytes and 14,000 leukocytes per cubic millimeter, with a differential of 80 per cent polymorphonuclear cells. The urine contained acetone and a faint trace of albumin. The centrifugalized urinary sediment showed, per high dry field, five to six white cells, five epithelial cells, fifteen

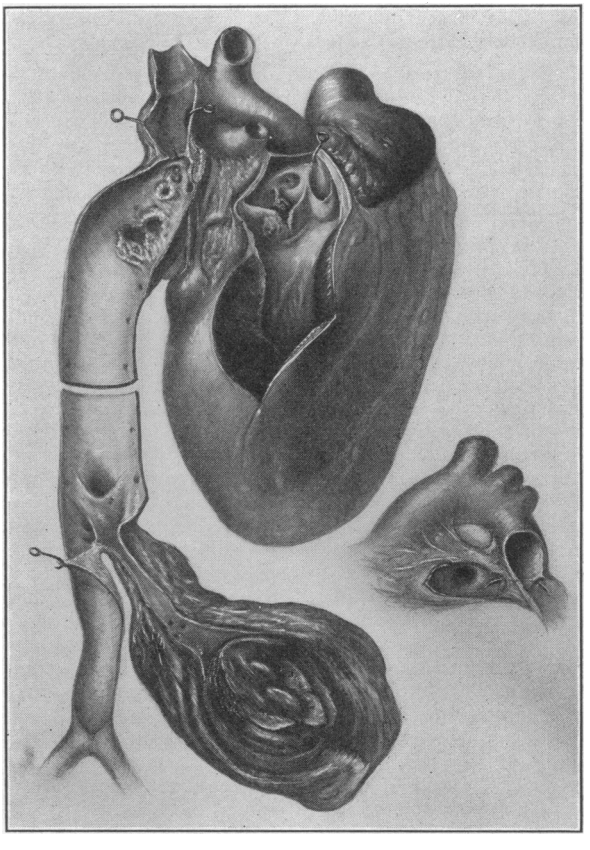

Fig. 1.—Drawing of heart, aorta and superior mesenteric artery. The vegetations on the aortic leaflets, the ruptured aortic valve, the bifid coronary mouth, the coarctation of the aorta, the patent ductus arteriosus, the mycotic aneurysm of the aorta and the mycotic aneurysm of the superior mesenteric artery are illustrated. The inset shows a bristle in the patent ductus arteriosus. The same bristle emerges from the ductus arteriosus just below the coarctation in the main figure.

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to twenty red cells, and one to two granular cellular and hyaline casts. The Wassermann reaction on the blood was negative.

Diagnosis and Treatment.—A diagnosis of partial intestinal obstruction was made and an exploratory laparotomy was performed the day of entry. Some peritoneal adhesions were found and freed, but a definite intestinal obstruction was not present. A fusiform swelling, approximately two centimeters in diameter, showing expansile pulsations, was seen in the region of the superior mesenteric artery. This was thought to be an aneurysm. Numerous enlarged lymph nodes were present in the mesentery.

Following the operation, the patient continued to have occasional attacks of generalized abdominal pain. The phenolsulphonephthalein excretion, the Mosenthal test, and the non-protein-nitrogen and creatinin determinations in the blood were within normal limits. The Wassermann reaction on the spinal fluid was negative. The urinary findings and leukocytosis continued as on entry, and the anemia became more marked. X-ray films of the renal regions showed no evidence of stone. A tentative diagnosis of subacute glomerular nephritis was made.

An examination eight days after entry showed no change in the size of the heart; the brachial blood pressure was 185 systolic, 105 diastolic. A loud systolic murmur was heard anteriorly in the third left interspace which was transmitted to the neck, left axilla, and back. This was thought to indicate a congenital cardiac lesion.

Three weeks after entry the patient was transferred to the Medical Service. At this time a diastolic murmur was heard to the left of the sternum in the third and fourth interspaces. The brachial blood pressure was 150 systolic, 60 diastolic. The next day definite aortic systolic and diastolic murmurs were heard, and a to-and-fro murmur was present over the pulmonic area. A very loud, rough, systolic murmur was heard over the posterior thorax, maximal along the spine. Slightly enlarged, tortuous, transverse scapular arteries could be easily felt. The radial pulse was Corrigan in type, and a pistol-shot sound could be heard over the brachial artery, but not over the femoral. Pulsation in the dorsalis pedis and internal malleolar arteries was imperceptible. The brachial blood pressure was 160 systolic, 25 diastolic; the femoral was 85 systolic, 70 diastolic.

Because of these findings, a diagnosis of coarctation of the aorta, aortic regurgitation, and probable patent ductus arteriosus was made. As a subacute bacterial endocarditis could explain the anemia, leukocytosis, urinary findings and the changing cardiac signs, cultures of the blood were taken from which *Streptococcus alpha* was obtained. The same organism predominated in a culture of the throat and was also found in the urine. A urea-clearance test was within normal limits.

X-ray films and fluoroscopy of the chest were normal, save that the aorta could not be traced beyond the transverse arch, though the vessels in the upper mediastinum pulsated abnormally. The rib margins were not scalloped. An electrocardiogram showed an intraventricular block and a right axis deviation. Simultaneous pulse tracings showed that the pulsation of the radial artery preceded that of the femoral (normally, the femoral pulsation precedes the radial).

The patient's temperature, heretofore practically normal, now became moderately elevated, and remained so until death. The arms continued to show a high pulse pressure with low diastolic pressure, and the legs a low pulse pressure with relatively high diastolic pressure. Dyspnea and orthopnea now appeared, soon followed by pulmonary edema and then a right pleural effusion. On the day of death a gallop rhythm and a pericardial friction rub were heard. The spleen was palpated for the first time, and the edge of the liver was found to be four centimeters below the right costal margin. He died of cardiac failure a month after entry. No petechiae were noted at any time.

The clinical diagnosis was coarctation of the aorta, patent ductus arteriosus, subacute bacterial endocarditis, rupture of the aortic valve with consequent cardiac failure and focal glomerular nephritis. The abdominal aneurysm was thought to be congenital.

Postmortem Findings.—Postmortem examination (performed three hours after death): In the conjunctiva of the lower left eyelid were two petechial hemorrhages. There was slight clubbing of the fingers of the right hand. The recent lower right rectus operative scar had a central granulating zone.

The peritoneal cavity contained a slightly increased amount of dark, straw-colored fluid, and in the right lower quadrant the peritoneum was iced over by a recent fibrinous exudate. Each pleural cavity contained 250 cubic centimeters of a light, straw-colored fluid. The pericardial sac, likewise, contained an increased amount of clear, straw-colored fluid. An old, thin, fibrous band joined the epicardium and pericardium at the apex.

The heart weighed 400 grams. The left ventricle was hypertrophied, having a maximum thickness of 1.7 centimeters. The aortic valve was partially destroyed by a large, crumbling vegetation, and appeared to be composed of two leaflets. The right cusp and the posterior cusp were fixed together by the vegetation, which had destroyed their common attachment. A portion of the right cusp had been torn out of this fibrinous mass. An anomalous bifid coronary artery was situated behind this cusp.

The aorta was constricted in an hour-glass fashion by a firm, white band 2 centimeters below the root of the left subclavian artery. This band, causing a coarctation, was less than 1 millimeter in thickness. Immediately distal to it the intima was puckered around a small perforation, through which a bristle could be passed into the pulmonary artery. This was the remnant of the ductus Botalli, which was anatomically open but functionally closed. Measurements of the circumference of the aorta were 1.9 centimeters at the bifurcation, 3 centimeters at the level of the superior mesenteric artery, 4 centimeters at a point 2 centimeters below the coarctation, 1.9 centimeters at the coarctation, and 2.8 centimeters at the root of the left subclavian artery. There were numerous large, irregular vegetations and small aneurysmal dilatations of a mycotic nature distal to the coarctation.

The branches of the arch of the aorta were normal in number and location, but were slightly larger than normal in size. The intima of this portion of the aorta showed a few yellowish, atheromatous streaks. At a point 5.5 centimeters from the aorta the superior mesenteric artery dilated to form a fusiform mass measuring 6 x 5 x 4 centimeters, situated in the mesentery of the small bowel. The walls of the artery were greatly thickened and held a laminated thrombus in the lumen. The vessel could not be traced beyond the aneurysm. The internal mammary arteries were larger than normal, but their anastomoses with the intercostal arteries were not unusual. The heart, aorta, and superior mesenteric aneurysm are shown in Figure 1.

The spleen weighed 550 grams, was soft in consistency and contained several small, firm, white, embolic abscesses. Each kidney was enlarged, showing purple petechiae beneath the capsule, and on section this "flea-bitten" appearance was still more striking.

Microscopic Findings.—The aortic vegetations were composed of clumps of Gram-positive organisms surrounded by dense fibrin. The mycotic aneurysms of the aorta showed a dense, infected, granulation tissue. The aneurysm of the superior mesenteric artery was similar, save that an organized and canalized thrombus was also present. The sections of the lung were examined for the changes usually ascribed to rheumatic

fever;[8] but none could be found. A recent terminal bronchopneumonia was present. The serosa of the jejunum was covered by a recently organized fibrinous exudate. The spleen showed numerous infected embolic abscesses situated in an acute, gray, splenic tumor. The typical picture of an acute focal glomerulonephritis was present in the kidneys.

Pathological Diagnosis

I. Congenital heart disease.
   1. Coarctation of the aorta.
   2. Anatomically patent ductus arteriosus.
   3. Bifid mouth of right coronary artery.
   4. Left ventricular cardiac hypertrophy.

II. 1. Subacute bacterial endocarditis (*Streptococcus alpha*). involving the aortic valve with rupture of the right cusp.
   2. Mycotic aneurysm of descending thoracic aorta.
   3. Mycotic aneurysm of superior mesenteric artery.
   4. Acute focal glomerulonephritis.
   5. Acute gray, splenic tumor with embolic abscesses.
   6. Passive congestion of the viscera and bilateral pleural effusion.

III. Old localized adhesive pericarditis.

IV. Recent localized peritonitis.

V. Terminal bronchopneumonia.

COMMENT

The incidence of bacterial endocarditis in a congenitally malformed heart is quite high. In this instance the oldest lesions appeared to be in the thoracic aorta near the coarctation, followed by involvement of the superior mesenteric artery and then the aortic valve. The onset of the infectious process probably occurred a year before entry when the patient was said to have had pneumonia. The process was fairly quiescent up to the onset of the symptoms before entry, and then became more fulminating. Death, however, was due to cardiac failure rather than to sepsis. Distortion and rupture of the aortic valve imposed too great a strain on an already overburdened cardiac muscle.

The symptoms during the terminal stages of the disease were difficult to interpret, and several diagnoses were considered before the correct one was reached. The first diagnosis of intestinal obstruction was based on the history of a ruptured appendix in childhood, and the abdominal colic and the vomiting. When the abdominal aneurysm was found, the blood and spinal fluid were examined to rule out syphilis. Acute hemorrhagic nephritis was next considered because of the hypertension, anemia, and hematuria. Finally, the probability of congenital heart disease with coarctation of the aorta was recognized, and the complication of subacute bacterial endocarditis suspected and confirmed by culture of the blood.

The coarctation was diagnosed because the arteries of the upper extremities showed hypertension and a high pulse pressure, as contrasted with a hypotension and low pulse pressure of the arteries of the lower extremities. Furthermore, the enlarged, tortuous, transverse scapular arteries were evidence of collateral circulation. The lack of scalloped rib margins in the x-ray films is not unusual at this age, as apparently only in older individuals who have had coarctation for a longer time does this sign appear. Likewise, the early arteriosclerotic changes found in the aorta would have advanced with time.

In the presence of hypertension in a young individual, coarctation of the aorta should always be considered, and the character of arterial pulsations in the lower extremities noted.

SUMMARY

1. A case of coarctation of the aorta with subacute bacterial endocarditis producing mycotic aneurysms of the aorta and the superior mesenteric artery is reported. The symptoms resulting from the aneurysm of the superior mesenteric artery simulated acute intestinal obstruction and a laparotomy was performed. The urinary findings actually due to the bacterial endocarditis led to a tentative diagnosis of acute hemorrhagic nephritis.

2. Coarctation was finally recognized because of the marked difference in arterial pulsations in the upper and lower extremities, and because of evidence of collateral arterial circulation.

3. The rapidly changing cardiac murmurs, anemia and hematuria led to the diagnosis of the complicating subacute bacterial endocarditis.

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SOME NEW INSTRUMENTS AND MODIFICATIONS

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FIGURE ONE.—One of the inconveniences of working in the vaginal canal has been the interference with vision by the handles of the several instruments used. In nose and throat work some of this has been obviated to a great extent by the development of the "bayonet" forceps.

In Figure 1 is shown a view of the ordinary cervical tenaculum, to which this principle has been applied. Its advantages are those of increasing the visibility of the operating field in that the handles, and incidentally the assistant's hands, are out of the field of vision; and secondly, that when one engages the cervix on each side, the increased leverage due to the angulation of the instruments brings the cervix down into the lower vaginal segment with greater ease and gentleness, that is, with less direct tractive force.

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Figure 2.—As a victim of three laparotomies, my memories have always been lacerated by the thoughts of the unconscious brutality of the usual silkworm-gut retention sutures—with their tend-